CLINICAL PROCEEDINGS

of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

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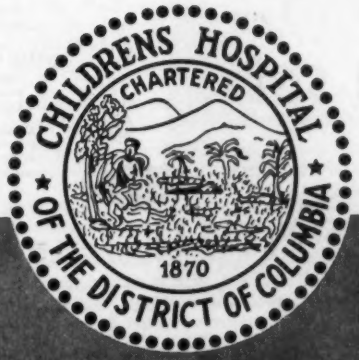

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OF THE CHILDREN'S HOSPITAL

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CONTENTS

MOTH BALL POISONING. *Robert Bregman, M.D.*..... 1

ENDOCARDIAL FIBROELASTOSIS. *John Nestor, M.D.*..... 3

SCHIZOPHRENIC-LIKE PSYCHOSES IN CHILDREN. *Belinda Straight, M.D., Sidney Berman, M.D., Reginald S. Lourie, M.D.*..... 6

CLINICO-PATHOLOGICAL CONFERENCE. *E. Clarence Rice, M.D., Roger Bergstrom, M.D., Leroy E. Hoeck, M.D.*.. 17

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MOTH BALL POISONING

A Case Presentation

Robert Bregman, M.D.

CASE REPORT

M. D.,* a three year old colored male was admitted to the Children's Hospital on September 5, 1953 because of vomiting and abdominal pain of two days duration. The pain was generalized and of moderate severity. He ate breakfast the day of admission, but refused the noon and evening meals. There was no other significant history.

It was noted in the dispensary that the child was very pale and close questioning revealed an adequate dietary history and denial of ingestion of any foreign substances.

On admission, the child was a well developed and well nourished colored male with pale mucous membranes and slightly icteric sclerae. He was somewhat listless but did not appear acutely ill. The liver edge was palpable two fingers breadth below the right costal margin.

Initial laboratory studies revealed a "dark" urine with acetone and white cell clumps. A benzidine test on the urine was negative. The hemogram was as follows: Hemoglobin 3.5 gms, hematocrit 17 per cent, rbc 1,300,000, wbc 27,400 with 69 per cent polys, 6 per cent bands, 3 per cent young forms, 21 per cent lymphocytes and 1 per cent eosinophile. Sickle cell studies on admission and two days after admission were negative. The reticulocyte count was 26.5 per cent. The platelets were normal and the peripheral smear had many nucleated red blood cells with anisocytosis and varying degrees of hypochromia. There were many fragmented and distorted red blood cells. Heinz bodies were described in the peripheral smear by one observer.

The hemogram was repeated in twenty four hours and was essentially unchanged. The Coombs test was negative and the erythrocyte fragility test showed increased resistance in hypotonic saline. VDRL serology test, cephalin flocculation, total protein and A/G ratio, serum bilirubin and prothrombin studies were normal. Urine and fecal urobilinogen levels were elevated and the stools were negative for ova and parasites.

Three days after admission, the mother reported that she remembered taking some mothballs out of the patient's mouth and hands three or four days before admission to the hospital.

The patient was transfused the day after admission and became very active and alert. His hemoglobin and hematocrit rose steadily and he was discharged six days after admission, with a hemoglobin of 9.5 grams, hematocrit of 36 per cent. His physical examination was normal. Prior to discharge, bone marrow examination showed marked erythroid hyperplasia and moderate eosinophilia.

DISCUSSION

Intoxication from moth balls containing naphthalene was first reported in the American literature in 1949, but had been previously described in Europe in 1899.

* On the service of Dr. Edward Lewis.

Naphthalene produces an acute severe hemolytic anemia which at times has been fatal. The hemolytic process is readily recognized, but this etiology is rarely suspected on admission to the hospital.

Moth ball ingestion is most common in children between two and four years of age, this group being more prone to put foreign objects in the mouth. This condition usually occurs in the spring and fall seasons when clothing is being taken out or put into storage and moth balls become common household items.

The usual route of intoxication is via the gastrointestinal tract. However, there are reports in foreign literature of poisoning due to inhalation of naphthalene vapors. Schafer, in 1951, reported a fatal case of acute hemolytic anemia in a newborn infant. This baby did well until he went home from the hospital. At that time diapers were used which had been stored in moth balls and which smelled strongly of moth balls even though they were rinsed prior to use. The infant's skin had been rubbed with olive oil and it was believed that the oil aided the absorption of naphthalene.

The first four cases reported in American literature were reported by Zuelzer and Apt in 1949. The diagnosis was based on the typical clinical picture of an acute hemolytic process plus the history of moth ball ingestion. In one of their four cases, naphthol was isolated in the urine. This is a derivative of naphthalene not found in normal urine.

After ingestion the naphthalene is absorbed in the gastrointestinal tract. Oxidation occurs and it is the oxidation products of naphthalene which are toxic and cause hemolysis.

The presenting symptoms vary and may be related to the upper respiratory, gastrointestinal or genitourinary systems. The usual presenting complaints are anorexia, vomiting, listlessness, or lethargy, pallor and wine colored urine.

Physical examination usually reveals an acutely ill child with fever, pallor and possibly mild jaundice. The liver and spleen if palpable are only slightly enlarged. An important diagnostic point is the odor of naphthalene in the urine and stools.

Laboratory studies reveal a severe anemia with hemoglobin in the range of 3.5 to 5.5 gms. and red blood cell counts of 1.5 to 2 million. There is usually a leucocytosis of from 10,000 to 40,000 and the plasma oxygen hemoglobin is elevated.

The peripheral smear reveals fragmentation and spherocytosis. The abnormal cells have sometimes appeared similar to sickle cells. However, the sickle cell preparation is negative. Heinz bodies will usually be seen if looked for. The Coombs test is negative and there is usually increased red blood cell fragility in hypotonic saline. There may be an increase in serum bilirubin and increased excretion of stool and urine urobilinogen if the blood

destruction has been rapid. Hemoglobinuria and albuminuria are often found and the urinary sediment usually contains white cell casts and occasional red blood cells. After hemolysis has occurred, the treatment of naphthalene poisoning is transfusion of whole blood. There is no specific therapy. If the patient is seen immediately after moth ball ingestion, the stomach should be lavaged in an effort to remove the toxic substances before absorption.

SUMMARY

This case is presented because moth balls containing naphthalene produce an acute hemolytic anemia which usually is very serious and may be fatal. The hemolytic process is readily recognized but the etiology is usually missed due to failure to think of this disease.

The disease is most common in the two to four year age group and is more likely to occur in the spring and fall seasons.

Moth ball poisoning should be suspected in any case of sudden hemolytic anemia in children. Parents should be very closely questioned regarding this possibility.

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ENDOCARDIAL FIBROELASTOSIS

John O. Nestor, M.D.

Endocardial fibroelastosis is a condition which occurs almost exclusively in infants and is characterized by a diffuse thickening of the endocardium plus hypertrophy, dilatation and failure of the heart. It has been described under a variety of names including fetal endocarditis, congenital idiopathic hypertrophy of the heart, endocardial fibrosis, prenatal fibroelastosis, elastic tissue hyperplasia, and endocardial sclerosis.

The etiology is unknown but a variety of theories attribute the condition to infection before or after birth or consider it a congenital defect either due to a genetic factor or a uterine environmental influence such as anoxia. Other possibilities are that it is a degenerative process, a metabolic disorder, or a collagen disease. It is one of the most common types of fatal heart disease in infants and occurs only slightly more frequently in males than

in females. The race distribution seems to be in proportion to the general population. It occurs almost exclusively in infants but is occasionally found in older children and is rarely reported in adults.

Bell, Pollack, and Rice recently reviewed 2672 autopsies reported at Children's Hospital, Washington, D. C. during the period from January 1935 to July 1953. They listed 196 cases of congenital defects of the heart in this series including 13 cases of endocardial fibroelastosis. Of these nine were under, and four were over a year of age, and eleven were either dead on arrival or were hospitalized less than 24 hours before death, and two lived longer than 24 hours in the hospital.

The signs and symptoms include marked cardiac enlargement, dyspnea, cough, tachycardia, irritability, anorexia, vomiting, and predominantly left sided heart failure with pulmonary rales, large liver, peripheral edema and variable cyanosis and murmurs.

There are three clinical types in this condition. Approximately a quarter of the patients have fulminating illnesses in that death occurs a few minutes or hours after onset, which is usually under six weeks of age. In about one half of the infants, survival after onset is usually less than four weeks and the onset is usually between six weeks and six months of age. In both of these, growth, development and nutrition are good. The remaining 25 per cent of the patients are usually older infants and children, and are characterized by slow onset, impaired nutrition, and retarded development. In one series the onset of symptoms was before two weeks of age, in one third of the patients and before six months in the remainder. Death occurred within two weeks of onset in 51 per cent and before two years of age in 91 per cent. In another series 92 per cent were dead by one year of age.

If the child survives long enough to allow the performance of laboratory procedures, the blood count is usually normal although occasionally there may be a secondary anemia. Leukocytosis only occurs where there is secondary infection. The sedimentation rate, serology, urine analysis, and tuberculin skin tests are all normal. X-ray and fluoroscopy reveal predominantly left sided cardiomegaly. The electrocardiogram is usually abnormal, showing evidence of an abnormal myocardium and left ventricular hypertrophy by means of low, diphasic or inverted T waves and depressed ST segments usually in leads 2, 3, AVF, V_5 and V_6. The conduction is usually normal.

The differential diagnosis includes coarctation of the aorta, subaortic stenosis, anomalous left coronary artery, glycogen storage disease of the heart, pericarditis with effusion, myocarditis, rhabdomyoma, beri beri, heart disease, acute nephritis with cardiac failure, other congenital malformation, and arteriosclerotic heart disease.

The treatment is that of cardiac failure of almost any other etiology but is usually of little avail in the fulminating and acute types but often prolongs life a few months or years in the chronic type.

The pathogenesis, like the etiology, is not yet established. One theory is that hypertrophy and dilatation of the myocardium results from stasis and anoxia occurring in the sub-endocardial myocardium when arterioluminal, arteriosinusoidal, and thebesian vessels have been obliterated by fibrosis of the endocardium. A more recent and more probable explanation postulated by Johnson is that endocardial fibroelastosis always develops in hearts having associated malformations which could cause endocardial anoxia such as anomalies of the coronary artery, premature closure of the foramen ovale, and valvular atresias.

The gross pathology is characterized by a thickened endocardium, predominantly left sided cardiac enlargement, hypertrophy, dilatation, and in about one half of the cases by valvular deformation characterized by thickened, nodular, wrinkled leaflets which are sometimes fused producing stenosis. There are no surface vegetations.

Microscopically there are increased amounts of collagenous tissue arranged in layers parallel to the myocardium and including large numbers of elastic fibers. The subendocardial-myocardial junction is not well defined and there are finger like projections of endocardium into the myocarduim with hypertrophy of the myocardial fibers and perhaps hydropic degeneration. No Aschoff bodies or cellular infiltration are present. Calcifications are common in the endocardium, myocardium, and papillary muscle.

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SCHIZOPHRENIC-LIKE PSYCHOSES IN CHILDREN

A Review of the Literature and Presentation of Two Cases

Belinda Straight, M.D.

Only a short time ago schizophrenic-like psychoses in children were considered rare occurrences. The outlook was thought to be hopeless. Now that this entity is widely accepted, pediatricians are more on the alert for danger signals. It will be they who will see the first relationship disturbances in these children. Early diagnosis and therapy remain vitally important. In view of this it was felt that a review of the literature and presentation of two cases were in order.

Criteria. Bender[1] describes schizophrenic children as revealing "pathology in behavior at every level and in every area of integration or patterning within the central nervous system, be it vegetative, motor, perceptual, intellectual, emotional or social. Furthermore, this pathology disturbs the pattern of every function in a characteristic way."

Bradley and Bowen[3] offer the following criteria for schizophrenia in children:

1. Seclusiveness.
2. Irritability when seclusiveness is disturbed.
3. Daydreaming.
4. Bizarre behavior.
5. Diminution in number of personal interests.
6. Regressive nature of personal interests.
7. Sensitivity to comment and criticism.
8. Physical inactivity.

Potter's criteria[13] are applicable to a greater number of patients and include:

1. A generalized retraction of interests from the environment.
2. Dereistic thinking, feeling and acting (thinking dominated by fantasy).
3. Disturbances of thought, manifested through blocking, symbolization, perseveration, incoherence and diminution, sometimes to the extent of mutism.
4. Defect in emotional rapport.
5. Diminution, rigidity, and distortion of affect.
6. Alterations of behavior with either an increase in motility leading to incessant activity or diminution of motility leading to complete immobility or bizarre behavior with a tendency to perserveration or stereotypy.

To these Despert[6] adds:

1. Excessive dependence on the mother.
2. Non-functional play.
3. Temper-tantrums.

Diagnosis. Diagnosis of such a serious condition should be made by a competent psychiatrist in conjunction with such psychological tests as can be utilized for the particular child.

Onset. The onset may be insidious or acute. In infantile autism* a mother may complain that her baby never has responded toward her in the way that previous children responded. Sometimes an acute episode in an outwardly normal child will follow hospitalization. The first years, the years from three and a half to four and a half, and the prepubertal ones are common periods for the development of schizophrenia[12].

Symptoms. The symptoms vary with the age of the child, his relationships in the family group, and the degree of deterioration. Of all the symptoms which may be found, most striking is the lack of emotional rapport with others. The give and take between two people has not been awakened or is nonfunctioning. The patient is dependent, excessively so, upon his mother. Yet he appears to be incapable of affection. He does not look one in the face. People become tools to fetch and carry for him. A typical remark is that of a little girl to her mother, "I can't use you now."

As a group they are withdrawn and angered by interruption of their lonely pursuits. The spontaneous, changing playfulness of the normal child is lacking. Speech (which depends on the desire to reach out for another) may not develop at all or may vanish, severing more completely the tenuous contact between child and parent.

Motility disturbances. Bender[2] in her study of 100 schizophrenic children at Bellevue Hospital describes vividly the characteristic motility. The patient tends to be awkward, has problems of balance, is hesitant about climbing stairs. He uses the balance of the grownup as his own. The center of gravity of the grownup becomes his center of gravity. He may be rocked, or wrapped around the body of the adult and he complies. When he is placed with feet together and arms out, one may turn his head and the whole body follows the head, a phenomenon which Bender called "whirling." This is not generally found in normal children past the age of five.

The compliance of the child's body is explained as being due to his inability to see himself as a whole person apart from others. He does not know where he begins and ends and where the other person begins and ends.

Autonomic nervous system. Changes in the autonomic nervous system have been described by Bender. These effect the flow and control of saliva. The temperature regulation is described as particularly labile.

Behavior changes. The pediatrician is often consulted because of feeding difficulties. The child (perhaps never a demanding baby for food) may be

*Autism is used here as the earliest possible manifestation of schizophrenia. Kanner points out that schizophrenia can and does begin as early as the diaper age.

indifferent to food at toddler age. He often shows difficulty in falling asleep. Patterns of sleep, previously normal, may change. In the beginning these may be the presenting symptoms. The physician will have to differentiate the normal sleep difficulties of the 18 month old child and the normal two year old anorexia from these more profound changes which are accompanied by emotional flattening and withdrawal. The desire and the postural anticipation of the normal child for being taken up in the arms of the mother is lacking in the autistic child and according to Kanner is an important sign. Behavior may lapse into the grossly bizarre, grimacing, hitting oneself, or hitting at hallucinatory figures. Repetitive actions, rituals, and compulsions are observed in some cases.

Emotional disturbances. The affect is rigid or flat. It may be grossly inappropriate, laughing suddenly while alone or hitting at the air. Noises from the outside, a door slamming, someone calling, will throw a child into a severe temper tantrum. All these represent interruptions of fantasy. It should be emphasized that the mildest frustration becomes unbearable to the schizophrenic child.

Anxiety is a prominent feature of such patients. Bender points out that anxiety in any child (not accompanied by a reality situation warranting such emotion) deserves serious investigation.

Social disturbances. Socially these patients may be appealing children. They appear so helpless and so anxious that they soon become favorites on the wards. Sought after by normal children, they are also puzzling to them. The schizophrenic-like child provokes anxiety in adults, partly because he cannot be reached by the ploys which delight other children.

We have mentioned excessive dependence on the mother and indifference to her. It may as well be someone else lifting, rocking, supplying food. These children have trouble in differentiating between people. A hand is taken to lead a little girl across a room. She does not care to whom the hand belongs, so long as she may use it to propel her across the floor. As she has difficulty differentiating herself from others, how much more difficult it must be to differentiate one person from another! One child described by Despert said (instead of using the word "Daddy"), "That man who sleeps here, has bacon and eggs in the morning, that man(4)."

Speech. Because the concept of self as distinguished from others (the ego) is lacking, the first and the third person singular are frequently confused. "Give you a candy" stands for "give me a candy." The schizophrenic child may continue to call himself "you" until eight or ten, when he will learn "I." The ordinary child will make that change by the age of two and a half or three.

Speech varies from lack of development and mutism after it has once been learned to a highly intellectualized pattern. Echolalia is another way

of taking on another person as part of oneself. Kanner[9] writes of delayed echolalia, in which a complicated word having no immediate interest to a child of that age will be stored in the memory and used long afterwards. Some of these children have phenomenal vocabularies; however words become an earnest pastime, a serious occupation without the sense of joyousness the ordinary five-year old might experience with words.

Intellectual growth. Hypomotility, physical dependence, and lack of speech development in an autistic child may cause him to be placed in an institution for the feeble-minded. On the ordinary IQ tests he may perform poorly because communication is faulty. The Rorschach will often help in the differential diagnosis, as would a history of normal development followed by regression.

Some schizophrenic children exhibit overgrowth intellectually in one direction. A one and a half year old child in Kanner's[9] series could identify eighteen symphonies and their composers. Some become fascinated by the collection in their memories of telephone or license numbers. Astrology can become a preoccupation for a six year old. Just as some of the speech and intellectual pursuits are abstract, removed from every day life, so also the art of these children tends to be intellectualized and abstract.

At the other end of the story is the child who though once competent in scholastic work has deteriorated to a level far below his chronological age. In such a child the prognosis is not favorable.

Case No. 1

T. D., a four and a half year old boy was brought to Children's Hospital for mutism and weight loss of 15 pounds within two weeks. He is the son of an unwed school teacher mother; the father was not disclosed. Delivery was normal. From a foundling home where he spent his first six months he went to a foster home where he has remained. He sat up at six months, stood at twelve months, walked at twenty-three months and talked in short sentences at two years. His foster mother had a baby of her own when Teddy was two and a half. At this time he stood in his bed screaming for a long period, refused solid foods and became mute. He withdrew in his daily contacts with people. Since that time he has been heard to speak only when alone playing, or during sleep. When he was four and a half, the foster mother had her second child. Teddy again had eating disturbances, withdrew markedly and had difficulty in falling asleep. Medical history was essentially negative.

On the ward he was noted to be a small, markedly cachetic (21 pound) child of 4½ years. He had frontal bossing and strabismus. He sat quietly at a table looking down. He fulfilled Kanner's description of silent wisdom. When food was placed before him he would look at it, then at his hands. But he would not initiate movement toward it until this was suggested. He clung to any adult who approached him, but did not prefer one to another, and would not look in their faces. He used the adult's hand to propel him from one side of the room to the other. He could be rocked in any direction, relying on the body of the examiner for his own balance. While rocking back and forth a secret smile played across his face. Occasionally he broke into wild peals of laughter. He did not whirl when his head was turned. He was awkward in

maneuvering steps. He touched toys but did not play with them. He became restless and unhappy if the doors of the room were closed, and ran frequently to them, happy when they were opened. His actions were repetitive and lacked spontaneity. He soiled and wet the bed without showing concern. He was able to carry out detailed instructions. When asked to "take off the pen top, turn it around and fit it on the opposite end" he carried out these instructions.

The electroencephalogram showed frequent bursts of high voltage four to six per second, irregular spike wave formations and positive spikes. No focus was noted. This was construed to be an abnormal electroencephalogram. Other laboratory tests including fasting blood sugar and stools for trypsin were noncontributory.

During his hospital stay he began to eat more spontaneously, gained six pounds within two weeks, and began to use the dolls and toys for play. When awakening from sedation for EEG he repeated simple words after the examiner. He smiled broadly during this time. When he returned to the ward and was put in bed he screamed loudly and remained restless and noisy. For several days he echoed single words. He was discharged to a convalescent home. Since his arrival he has become mute again, but continues to show more interest in relationships than he had previously.

Case No. 2

Robert Q. was brought to the hospital when he was three years old by his mother who complained, "My son seems queer. He hollers. He bumps his head on chairs and tables. He slaps himself. He does not talk. He likes to stay in his crib. He refuses to go out of doors. He won't play with other children. There is no bladder or bowel training."

Robert was the third child of a college graduate who was frustrated in her attempt to become a librarian by the birth of her children. To Robert's arrival she had resigned herself without enthusiasm. To the father who spent much of the family savings on gambling, the son was something less than an asset, and he constantly questioned his own paternity. Three months after Robert's birth, husband and wife separated and the mother struggled through the year. When Robert was one year old Mrs. Q. noticed he was not interested in toys or herself. She might as well not have been present except at meal times. She got nowhere in bowel training.

Medical history: Measles at one and a half, chicken pox at two and a half, otherwise negative. Physical examination was essentially negative.

Developmental history: Weighed nine pounds at birth. Delivery was normal. He sat up at nine months and walked at one and a half years. He said only a word or two at two years; had temper tantrums when taken out of doors at two years. At three years he still refused food except from the bottle and particularly disliked colored foods in the bottle. Mrs. Q.'s big problem in the third year was controlling frequent nose bleeds which resulted from headbanging. When he was seen by the psychiatrist he banged his head against the floor and against his arm as he lay on the floor; he smeared saliva on the walls and whined or screamed. He was uncomfortable on physical contact but could not resist whirling. The diagnosis of autism was made and it was felt that under proper care the child might emerge. Robert was placed in a convalescent home and later a warm foster home was found. Mrs. Q. resisted Robert's removal at first, later resisted his return to her.

Robert cried and remained in his crib for three days after arrival at the foster home. The foster mother fed him from a bottle, comforted him in his restless sleep. Soon he began to join the family for meals. Gradually he accepted solid foods, showed

an interest in and dependency on the foster family, especially attaching himself to an older daughter. After two years with the foster family, and regular consultation and evaluation therapy, the child has gone through the hand to mouth phase and is interested in toilet training and succeeding in this area. He is gesticulating and imitating vocalizations to a limited extent. He still loves to rock. His play involves rhythmic toys and musical instruments. He responds to praise. He has emerged from a period in which he continually surveyed the playroom through the window of a tiny toy car, into a more open interest in others. He has begun to slap the observer instead of himself and shows interest in playing with children.

These two cases have been presented as two examples of the schizophrenic syndrome. In the first case of Teddy in which there is a large organic factor we feel that the emotional also plays an important role. We see difficulty in distinguishing human beings, speech disorders, sleeping and feeding troubles, excessive dependency and hypomotility, all following a traumatic episode on the birth of the foster child. We would like to emphasize that the schizophrenic syndrome can occur in any child, whether he has organic brain damage, is mentally retarded or is of normal capacity. One of these does not rule out the presence of the other.

In the second case we see a child who does not have a history of an abrupt onset, but already at one year seems different in his response to the world about him. Robert is now five, and is beginning to relate to people about him, having gone through the very early stages of development at a very late age. How far he will continue to grow is still uncertain.

Etiology. The etiology of schizophrenia is mostly speculative. Constitutional predisposition may be a factor as it is in adult schizophrenia. However, not enough cases have been gathered to warrant a positive statement in this direction.

In Kanner's[9] series the parents have all been intellectuals and professionals. Teachers, lawyers, physicians and psychiatrists make up the list. Many of the marriages are noted to be "cold and formal affairs." He found the mothers to be domineering, rejecting and lacking in spontaneous warmth. Despert[7] quotes a mother, "I can't stand my husband around the child," and "I'm going to raise me a nice child."

Rank[15] describes such a mother as having the ambition of being a perfect mother, a perfect wife, a perfect woman. She hopes through her child to find emotions which she is incapable of feeling herself. The child is made to carry a severe burden at an early age to supply the mother with emotions of which neither child nor mother are capable.

Tietze points out that the mother is remote, puts excessive pressure on the growing baby to achieve feats of development which are considerably beyond his age level. It is interesting to read several of the journals by parents of schizophrenic children about their children. They contain much objective data often sounding as if they had been written by an impersonal

onlooker. Most observers agree that the mothers have great anxiety and guilt about their schizophrenic children. The child's behavior alarms and bewilders them. We do not mean to imply that their anxiety has caused the child to be withdrawn, but rather that the child's behavior alarms the mother.

Whatever has happened to these children it is clear that a tremendous fear has driven them back or prevented their emergence from a state of primary narcissism. They lack trust in others and cannot identify with them. Thus normal development is shut off from them.

Therapy. Various types of therapy have included electroshock and insulin shock which do not appear to be the answer. Removal of the child to an institution set up to tolerate and understand such patients may occasionally be warranted. Other times a patient may be temporarily placed in a warm accepting foster home with psychotherapy on an ambulatory basis.

Rank, Despert[6] and Fabian[8] adhere to keeping the child at home and not making a further separation. Fabian feels the prognosis is poor for therapy in a schizophrenic child without treatment of the parent and advises treatment of the child and mother simultaneously. Treatment is aimed at helping the child relate to an adult and at helping the parent to consolidate these gains at home. The James Jackson Putnam Children's Center* is set up to treat schizophrenic children and their parents as well as providing nursery school. Such a situation is an ideal set-up not available is most communities.

Prognosis. The prognosis, formerly thought to be hopeless, is still very guarded. Lurie, Tietze and Hertzman[11] in ten cases found only one child making reasonable adjustment in the community. Potter and Klein[14] in fourteen cases had only one making a good adjustment. Cottingham[4] reports improvement in a case. Lourie, Pacella and Pietrowski[10], studying 20 patients over an average of eight years found improvement in nine, four having normal adjustment to the community. Lourie felt the prognosis to be more favorable in those children with an insidious onset than in those children with an acute onset early in life. We would also consider that the sooner treatment is begun, the better the prognosis.

DISCUSSION

Sidney Berman, M.D.†

On being asked to discuss the etiology of schizophrenia, it is difficult to make any comments without immediately touching on the mental health aspects of the problem and upon factors related to the diagnosis, prognosis and treatment of schizophrenia.

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Schizophrenia is a disorder which is widespread and presents a serious mental health problem. Approximately one-fourth* of all the hospital beds in this country are occupied by patients suffering from this psychosis. This does not take into account the many ambulatory cases which are never hospitalized and those which are never diagnosed. The factors creating this disorder are enigmatic. Many believe that the cause of this illness will be understood through the investigation of constitutional deficiencies. To date, such studies have been unrewarding in terms of explaining the nature of the illness or in its treatment. Others hold the opinion that a better understanding of the psychological, social and sociological factors may explain the nature of this disorder. That is, given the historical data in the life of the patient and his physical and psychological status, the illness can be understood as a specific reaction to stress situations in early life experience. Such a theoretical approach to the understanding of schizophrenia of childhood would place the pediatrician in the unique position of being able to detect early such disorders and may permit the pediatrician to support, in a preventative way, the parents in applying mental health principles to child-rearing.

Continuing this line of thought about constitutional and environmental factors precipitating this disorder, there are those who feel that the term *dementia praecox* should be used for the more constitutionally determined process and that the term *schizophrenia* should be used where the etiology seems understandable in terms of environmental factors and the course appears more benign. This would make it seem that the bizarre character of the symptoms and the intensity of the disorder would determine whether it was constitutional or functional. Such an approach could pose a danger in making a formulation based on descriptive evaluation rather than on an analysis of the dynamic process. In fact, the striking presentation of secondary symptoms such as hallucinations, delusions, and bizarre regressive behavior may overshadow what appears to be more fundamental to the nature of the disorder.

The primary basic disturbances are manifested in interpersonal relationships by a rigidity of affect, a self-preoccupation, a distorted reaction to reality or the world about the child, and a confused or incomprehensible language communication. This fits the pattern of the autistic child as described by Kanner.

Another unique characteristic of this syndrome is the disturbed mother-child relationship. If it is contended that the child's reactions are constitutionally determined as the basis for this disorder, then it could be understood how a mother's bewildered response and anxiety to such a child would tend to aggravate the situation. However, the investigation of

* National Institutes of Health, Bethesda, Maryland.

mothers of schizophrenic children shows that they have a strong hostility to the world about them and toward the rejected child. They appear to possess a basic coldness and an absence of easy relatedness and warmth to other human beings. Any physician who has had the experience of being with such mothers could easily recognize his own anxiety and his lack of ease in her presence which in turn could interfere with an objective relationship. If the primitive instinctual needs of the child are not adequately met by the mothering one, including a warm emotional symbiosis and instead, the child is subject to intense anxiety and unrelatedness by the mother, the only defense the child can experience in order to contain the anxiety will be by withdrawal. Such a withdrawal from the primary object relationship can be carried over to all other human contacts.

It is the function of the mother to be responsive to the needs of the infant. As the infant develops, this relationship facilitates the infant's space, time and reality development and socialization. If the child lacks such a relationship to the mother, it seems the child will be impaired in his relationship to other human beings too and therefore, able to deal with the world only in chaotic infantile ways. I think it is an amazing phenomenon how most mothers can pick up the right signal as to what is distressing a baby often even better than the physician. However, some mothers are completely lacking in such skills and are preoccupied with their own anxieties. Schizophrenia could be conceived as a reaction to living which does not occur over night but which gradually develops as a reaction to stress situations.

In the two children presented in this conference by Dr. Straight we are impressed by the absence of meaningful mother-child relationships and the infantile withdrawal and remoteness of their responses. Even if we were to grant that both children were constitutionally impaired, the psychological approach to the problem would still be the same. Therapy would be directed to the development of a primary relationship with a significant person to permit expansion of their relationships with others. This would hold even for the first case in which there also appears to be organic impairment. This case points out that regardless of the presence of other pathological factors, it is possible to develop a schizophrenic reaction as a defense against stressful life experiences. When both youngsters were brought into the room during Dr. Straight's case presentations, I am sure most of us could feel their unrelatedness to us, causing us to respond in kind, thereby making any sustained contact with them difficult. This, I feel, is one of the important diagnostic signs. Nevertheless, in such cases, it is important to rule out deafness or gross severe organic brain damage which also impairs a child's adjustment to reality. Returning to these two children, both of them had no consistent maternal relationships, were

subject to emotional deprivation and therefore psychologically adapted in primitive infantile ways to their life situations.

It would seem better to approach the problem of both children not in terms of therapeutic ambitions which could discourage any one of us and cause us to feel "what is the use." Instead we could try to understand what is happening organically and psychologically. When you have done that, therapeutics will take care of itself. With such a theoretical approach treatment at best takes a long time and requires a unique therapist to facilitate the child's emotional growth from primitive outbursts of hatred, confused communication and distrust to freedom from alarm, trustworthiness, and a belief in other human beings which permits personality expansion.

DISCUSSION

Reginald Lourie, M.D.*

It is important to stress the point made by Dr. Straight that schizophrenia in childhood is a response of the personality which is possible under different circumstances and in different types of children. In other words, it is possible for this type of personality reaction to be present in children with organic disturbances such as brain injuries and in children with mental deficiency, as well as children with none of these stigmata. It is only too easy when we see a schizophrenic-like response in a brain damaged child to blame the personality change on the anatomical changes.

It is also possible for these personality patterns to be of varying degrees. The cases reported represent relatively severe, long standing symptom-complexes. However, we see many instances of as severe changes of short duration. The most transitory is the familiar sight on the wards in the form of a severe withdrawal pattern; the child keeping out of contact with its environment in addition to regressive patterns reminiscent of infantile behavior. This type clears up within a few days of admission as a child adjusts to the ward or goes home. Such responses are on a situational basis and changing the situation clears it up. We can feel that the underlying process was relatively superficial. In other words, schizophrenic-like responses can be of varying intensity and depths. Some responses of this nature may be present for varying lengths of time from a month to a year and then clear up. We would have to suspect that these instances represent relatively superficial changes which have not "dug in." The cases reported here have been of sufficiently long standing to present more profound psychopathological changes so that even if recovery takes place it is not possible to know how far back on the road to normal levels these children can come. "Partial" recovery is possible.

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That there is a constitutional basis for the schizophrenic response, has long been suspected though as yet not demonstrated. Other children in the same family exposed to the same situations do not necessarily respond with the same reactions as the schizophrenic child. Some children present schizophrenic-like patterns from birth. If we think of the child's regression to much earlier patterns of behavior and the profound changes in relationship as basically a response to feelings of anxiety and the idea of being in danger, certainly there is a difference in the ability of the schizophrenic children to tolerate such feelings and this is probably thought of as being a constitutional factor. There is also a quantitative factor involved; that is the amount of anxiety and feelings of being in danger the child encounters must be considered.

In terms of other etiological factors, it is found that by far the most frequent disturbance is in the mother-child relationship patterns (see also Dr. Berman's discussion). We have to keep in mind that the anxiety experienced by the child can stem not only from not enough warmth, love, trust or security, but also from too much stimulation. The organism seems to be unable in some children to absorb large doses of pressure, tension or too much pleasurable excitation of a type for which it is not yet ready.

When the child retreats in the face of intolerable anxiety to more comfortable levels of personality development, this withdrawal is usually quite uneven with islands of intact function often left unmodified or even hypertrophied. This is only one puzzling aspect of these children about whom we still know so little.

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CLINICO-PATHOLOGICAL CONFERENCE

Directed by: E. Clarence Rice, M.D.

Assisted by: Roger Bergstrom, M.D.

By Invitation: LeRoy E. Hoeck, M.D.

The patient, a two weeks old white female, was born at term. The birth weight was six pounds three ounces. The baby was delivered by Caesarian section because of the mother's past history of cephalo-pelvic disproportion. At birth, the baby was noted to have multiple congenital anomalies and the mother was told the child would not live. The baby had presented a persistent feeding problem since birth and would take only one to two ounces at a time. The patient would vomit when she was fed more frequently than every four hours. The child had no bowel movements for two days prior to admission. The urine was dark in color.

Both of the parents and four siblings are alive and well. The mother had varicella during childhood and during the fourth month of pregnancy. The mother had all of her children by Caesarian section. A questionable miscarriage may have occurred at four months.

The past history was non-contributory.

Physical examination revealed a small, poorly developed white female weighing five pounds four ounces and appearing lethargic and weak. There was moderate dehydration and evidence of weight loss. The anterior fontanelle had nearly closed and the child had a receding chin. The circumference of the skull was 29.5 cm. and the cranio-heel length was 45 cm. The pupils were constricted and there were opacities in the lenses. The helix of the ears was redundant and pushed forward. The lips and tongue were dry and there was a partial cleft palate of the distal one-third. The chest measured 28 cm. in circumference and was clear to percussion and auscultation. No heart murmurs were heard. The liver was at the costal margin and the spleen was not palpable. The umbilicus was well healed. The skin was dry and scaly with evidence of weight loss. The skin had a slight icteric tinge and the child's appearance was described as "plethoric." There were six fingers on each hand, syndactyly of the second and third toes of each foot and swelling of the dorsum of the feet was noted. The Moro and sucking reflexes were poor.

Laboratory: On admission the hemoglobin was 14.3 gms.; the leukocyte count was 26,000 with 45 per cent segmented, 10 per cent bands, 3 per cent young forms, 27 per cent lymphocytes, 6 per cent monocytes and 9 per cent eosinophiles. The bleeding and clotting times were normal at this time. The hemoglobin and white cell count averaged 9.0 gms. and 50,000 respectively throughout the hospital course. Two weeks after admission however, the

hemolgobin had dropped to 4.0 gms. The differential remained essentially unchanged. The thymol turbidity and total serum bilirubin tests revealed a progressive increase from the time of admission with the former being 3.6 units and the latter being 8.0 mg./100 ml. on the last examination. Fragility test was negative. Bone marrow examination revealed marked stimulation of all elements. The urine was positive for bilirubin and urobilinogen on three examinations and many epithelial cells and bacteria were reported on two examinations. On one examination of the stool the urobilinogen was .25 mg./100 ml. of feces and the benzidine test was positive. The remainder of the stools were negative for urobilinogen. Chest x-ray revealed the cardiac silhouette to be somewhat widened to the right and to the left with no evidence of pathology in the parenchyma. Gastrointestinal studies gave an appearance suggestive of an incomplete obstruction in the region of the duodenum. The metaphysis of all the long bones showed demineralization and were atrophic in appearance.

Hospital Course: The patient continued to be a feeding problem and had repeated clay-colored stools with rather dark urine. The baby was placed on a hypo-allergenic diet and seemed to retain feedings better with slight weight gain. There was a bleeding episode from needle sites where a transfusion was given. Ecchymoses also developed about other needle sites. At no time was there a fever of any significance during the hospital course. Feedings continued to be regurgitated and were associated with weight loss and an increasing jaundice. Respiratory distress and cyanosis were marked and death occurred on the forty-ninth day of hospitalization.

STUDENTS' DIAGNOSES

Two of the students suggested mucoviscoidosis, and the rest suggested congenital biliary tract obstruction or atresia.

DISCUSSION

LeRoy E. Hoeck, M.D.*

This patient presents such a multiplicity of symptoms and signs that it seems as though no matter where one begins, whether it be with the general appearance including head, body and extremities, or with a consideration of separate systems, there is in any instance something abnormal to contend with. We know that congenital anomalies have always been an important factor in the fetal mortality picture and more so when compared with decreasing mortality from most other causes. Many anomalies are survived by the fetus in utero because the placenta has assumed the various functions that will be taken over by the organism itself immediately after birth. Waste products from the kidney, liver and other organs have

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been removed by the placenta, but a defect in the development of these organs must result in abnormal function after birth and ultimately in death when such a function is a vital one and is severely altered.

This two week old infant, who weighed four pounds four ounces on admission and six pounds three ounces at birth, has had a 15 per cent weight loss. Ordinarily a five to ten per cent loss might be expected before the first week but by two weeks this has usually been regained and the baby is back to better than his birth weight. With the history of taking feedings poorly in small amounts and vomiting, this loss can be accounted for on the basis of low fluid and caloric intake.

The history that the mother had chicken pox during her pregnancy would suggest that anomalies which were grossly evident might be due to a virus effect on the fetus, but we know that the fourth month of gestation probably is too late to produce these changes. Usually this is from an effect on the embryo during the first two months. Histories when looked at in retrospect can have wide margins of error, and although varicella has not been shown to have consistently deleterious effects on the embryo, there are some instances of mothers with varicella in early pregnancy who had babies with congenital defects. A virus infection such as rubella which was mild or inapparent in the mother during the first two months of pregnancy is another possibility. The history of four Caesarean sections and one miscarriage brings to mind the trauma and scarring that may accompany repeated sections and which might ultimately lead to an implantation of the placenta with deficient metabolic supply to the fetus.

The physical examination brings out some positive findings and some which are equivocal. The general appearance with moderate dehydration is compatible with the weight loss. The fine desquamation that is mentioned is not abnormal and often occurs in the neonatal period up to two or three weeks. The head circumference of 29.5 cm. is slightly larger than the chest and is within proper size relation to the chest at this age. When the head circumference is compared with the length by a ratio that is frequently used (viz. one-half the length in cm. plus 10 should equal the head circumference) we find one-half of 45 plus 10 equals 32.5 which is larger by 3 cm. than the head circumference. The fontanel is quite small at a time when it should actually be of good size. Compression and molding often cause a fontanel to be small immediately after birth, but as the days pass and the head takes on its normal contour, the fontanel becomes open and easily felt, and should be larger than the fontanel termed here as nearly closed. The eyes showed opacities in the lens. A fairly common occurrence of cataracts is in infants whose mothers have had rubella in the first trimester, typically with other congenital defects such as microcephaly, congenital heart disease and deafness in various combinations,

but we also see congenital cataracts alone, and in later months cataracts may be seen in mongolism and in galactosemia. As regards redundant ears, these are not infrequently associated with other anomalies, and again are mildly involved in mongolism. In renal agenesis the ears are quite striking, being low-set and without cartilage, but these infants die very quickly even before renal insufficiency can be responsible. The cleft palate may exist alone or it may be an accompaniment of other anomalies, and the receding chin and cleft palate are occasionally associated as an entity. Supernumerary fingers and syndactyly of the toes seems to be a part of the whole picture of multiple deformities, in this case without any familial background.

Jaundice can probably be discussed better along with the laboratory findings. It can be noted before going on to these findings that there was no evidence of umbilical infection, the chest was clear, x-ray showed evidence of enlargement of the heart, the liver was not enlarged and the spleen was not palpable.

The first laboratory value noted is 14.3 grams of hemoglobin, which is fairly good in an infant of two weeks. The next hemoglobin two weeks later however is four grams. A ten gram loss in this time would require quite a severe process. Bleeding could account for such a drop but there is little evidence of extensive bleeding at any internal site, and bleeding from superficial sites usually is not in great quantity and the quantity of blood loss in the bleeding episode mentioned here is not estimated. The hemoglobinemia in this instance was corrected to nine grams by transfusions.

Jaundice at this particular period has a number of causes. Jaundice with anemia could very well be caused by erythroblastosis, but the onset of the jaundice would have been much earlier, often within a few hours and usually within the first day after birth. Sepsis, with jaundice and anemia, would be another way to explain the generalized symptoms. The laboratory signs relative to explaining the jaundice do not fit together very well. The urine is said to be dark and when tested was positive for bile and urobilinogen. The stools are clay-colored and were negative for urobilinogen. Where the urobilinogen is coming from to give positive urine tests is difficult to explain as presumably the bulk of the urobilinogen is formed in the bowel tract, then goes to the liver and if not excreted there, gets into the circulation to be excreted by the kidney. On the other hand the presence of clay-colored stools, with bilirubin in the urine and not in the stool makes it logical to think that there is an obstruction to the flow of bile. Since it is not stated whether eight mg. of bilirubin was predominantly direct or indirect reading, this does not give us any additional help in determining whether the jaundice is obstructive or hemolytic.

The white blood cell count is another laboratory value that becomes difficult to interpret in the light of other findings. With the 25,000 white count subsequently going to 50,000 and being maintained there, what can cause such a high white count? An infection would be a logical conclusion, although one would suspect that sepsis with jaundice and anemia, if it is not responding to antibiotics, would overcome this baby sooner than it has. It is also known that a liver with congenital atresia of the bile ducts and obstruction and cirrhosis sometimes is accompanied by increased white counts, but the reason is obscure. The anemia and blood loss are sufficient reasons for stimulation of bone marrow elements. X-ray of the chest showing an enlarged cardiac shadow to each side would be compatible with a congenital heart and the suggestion by x-ray of duodenal obstruction would have to be a partial obstruction as stools are being passed.

To review the findings briefly in this patient, there are lens opacities and anomalous digits. The fontanel is very small, the head circumference equivocally small, possibly microcephalic. The combination of receding chin or hypoplastic mandible with cleft palate and glossoptosis is sometimes an entity in itself as described by Pierre Robin as a syndrome in 1929. Such infants nurse poorly and if the defect is severe, measures to keep the tongue forward, to lengthen the mandible, or provide a clear airway by tracheotomy are sometimes necessary. The x-ray findings of enlarged heart and an obstruction in the duodenum are suggestive of anomalies but not positive. The findings relative to the liver and bile ducts, with clay colored stools and bile in the urine speak for a congenital obliteration of the bile ducts as the primary factor in the jaundice, although when one comes down to the end stages, it would not be surprising to find the terminal cause of death was infection. It might be mentioned that a lesion that could account for partial intestinal obstruction and at the same time involvement of the extrahepatic ducts would be an annular head of the pancreas, but it is mentioned merely as a remote possibility.

PATHOLOGY REPORT

Roger Bergstrom, M.D.

At autopsy the body was that of an emaciated, severely jaundiced white female, weighing 2.49 kg., and measuring 45 cm. in length. The head was smaller than usual, the sclerae were markedly icteric. The following developmental anomalies were observed: cleft palate, six fingers on each hand and webbing between the second and third toes of each foot. The skin was dry and the turgor was poor.

All of the organs were jaundiced. The heart and great vessels were normal. The lungs showed scattered areas of atelectasis and the mucosa of

the bronchi were yellow. The tissue was crepitant. The liver was of normal size, the surface being smooth and dark green. It sectioned with resistance revealing a homogeneous dark green surface with indistinct lobulations. The gallbladder was small and pinkish-white in color and contained 4–5 ml. of white bile with a slight yellow tinge which could be expressed through the ampulla of Vater. The hepatic duct could not be demonstrated. The spleen and pancreas appeared to be normal as did the gastro-intestinal tract except for small petechiae which were scattered throughout the mucosa. The adrenals had a greenish-gray color and were somewhat enlarged. Their cut surfaces bled more than is usual. The right kidney was of normal size but the left was enlarged to twice its usual size, weight 34.5 gm. (normal 15 gm.) Capsules stripped with moderate resistance, revealing fetal lobulations and many small abscesses averaging 1–2 mm. in diameter on the surface. The sectioned surface showed similar abscesses containing green pus scattered throughout the parenchyma. The cortico-medullary junction was edematous and hemorrhagic. No other abnormalities were observed.

Microscopic examination of the lungs showed evidence of aspiration of foreign material and hemorrhage into the alveoli. There was marked hepatic fatty metamorphosis with distension of bile capillaries with bile pigment and a diffuse infiltration with leukocytes. The spleen and pancreas were normal. Moderate adrenal medullary congestion was present. The kidneys were congested and multiple abscesses were scattered throughout. A number of the glomeruli exhibited proliferation of the tufts, some showing accumulation of leukocytes. A few of the tubules were dilated and the tubular epithelium showed cloudy swelling with loss of nuclei. The bladder wall was infiltrated with leukocytes and there was some exfoliation of the epithelial lining.

Necropsy bacteriology:

Heart's blood culture—Pseudomonas sp.
Spleen culture—no growth.
Kidney, smear of pus—numerous gram negative bacilli.

Pathological Diagnosis:

1. Hemolytic anemia (clinical).
2. Bacteremia (clinical).
3. Multiple abscesses of kidneys.
4. Acute cystitis.
5. Intrahepatic biliary obstruction and fatty metamorphosis of the liver.
6. Atrophy of the hepatic duct.
7. Cataracts, bilateral.

8. Developmental anomalies:
   a. Polydactylism.
   b. Syndactylism.
   c. Cleft palate.

DISCUSSION

E. Clarence Rice, M.D.

One reason we selected this case was because the patient represented a diagnostic problem which is of considerable importance to the pediatrician. Because of the lack of fever and the increasing jaundice it was quite difficult to determine the etiology of the latter. In view of the leukocytosis and other blood findings I felt quite sure that we were dealing with an infection which it seemed was probably causing hemolytic anemia. I was somewhat surprised to find less evidence of obstruction to the biliary radicals when we examined the tissue microscopically than we expected.

The absence of fever in an infection in a baby or an elderly person is not too unusual. We have seen a number of babies who have had severe infections without any rise in temperature. It is interesting to note that despite the infection in the kidney the urinalysis apparently showed nothing abnormal. Occasionally one sees this. The parenchyma of the kidney is involved and does not communicate with the tubules and therefore does not allow pus to appear in the urine. In view of the pathologist's inability to identify the hepatic duct, one wonders if the atrophy was due to the intrahepatic obstruction of the biliary radicals resulting from the hemolytic anemia or whether the atrophy preceded this.

ANNOUNCEMENTS

Dr. Daniel Feriozi has resumed the practice of pediatrics with offices at 2173 North Glebe Road, Arlington, Virginia, following a tour of military duty as a medical officer with the United States Marine Corps at Camp Lejeune, North Carolina.

Professor A. Laurinsich, Relator in the next National Pediatrics Congress of the report "Disprotydemie in Infancy" for the purpose of collecting as complete a literature on this subject as is possible, would be very grateful to anyone working in this field for any copies of publications or even references which they could kindly send him.

Please address copies as follows:

A. Laurinsich, Clinica Pediatrica Universita, Parma, Italy

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